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Teaching Neuro*Images*: Dyspnea as a presenting manifestation of amyloid myopathy

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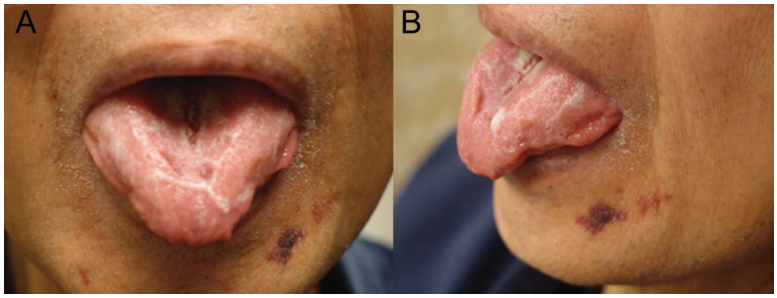
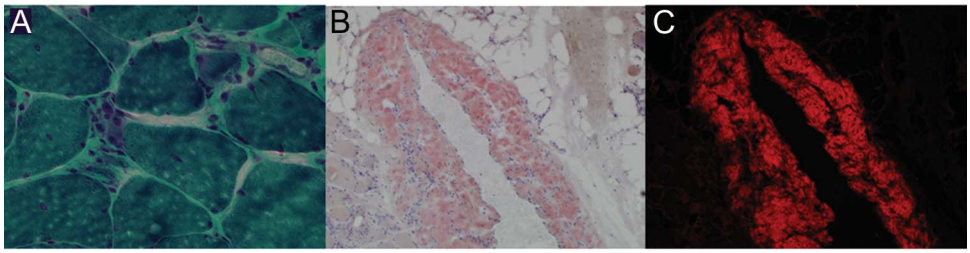
Figure 1 Face photograph showing macroglossia



(A) Front view and (B) side view.

Figure 2 Frozen sections of the deltoid muscle



(A) Trichrome stain shows a necrotic muscle fiber. (B) Congo red stain shows extensive amyloid deposition in a perimysial blood vessel wall that is intensely red when viewed under rhodamine optics (C).

A 69-year-old man had dyspnea followed by slowly progressive proximal leg weakness over 2 years. He had macroglossia (figure 1). Creatine kinase was 1,378 U/L. A deltoid biopsy revealed myopathy, denervation atrophy, and congophilic deposits around perimysial vessels, indicating amyloid (figure 2). Further workup revealed serum monoclonal lambda protein, bone marrow amyloid, and cardiomyopathy. Amyloid myopathy, an underrecognized entity, predominantly presents with progressive proximal weakness in primary amyloidosis.[1] Dyspnea results from cardiomyopathy or respiratory muscle weakness (our patient had both). Macroglossia due to amyloid deposition is a helpful clinical clue. The patient is on chemotherapy with cyclophosphamide, dexamethasone, and bortezomib, which improves prognosis in amyloidosis.[2]

AUTHOR CONTRIBUTION

Partha S. Ghosh: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and will give final approval. Greg M. Thaera: analysis or interpretation of data, accepts responsibility for conduct of research and will give final approval, acquisition of data. Jennifer A. Tracy: drafting/revising the manuscript, accepts responsibility for conduct of research and will give final approval, acquisition of data.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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2. Venner CP, Lane T, Foard D, et al. Cyclophosphamide, bortezomib, and dexamethasone therapy in AL amyloidosis is associated with high clonal response rates and prolonged progression-free survival. Blood 2012;119:4387–4390.

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OLFACTORY DYSFUNCTION IN PATIENTS WITH IDIOPATHIC INTRACRANIAL HYPERTENSION

Kah Fang Khoo, Bayan Lepas, Malaysia: This interesting pilot study[1] provides new evidence that patients with idiopathic intracranial hypertension (IIH) may have olfactory dysfunction. This feature is added to more conventional symptoms like headache, nausea, vomiting, and double vision. This is helpful as the Sniffin' Sticks procedure is less invasive than lumbar puncture and less expensive than MRI. Schmidt et al.[2] demonstrated reduction of olfactory bulb volume using MRI. A future study should include reduction of olfactory function as one of the minor criteria for diagnosing IIH and rule out space-occupying lesions and high intracranial pressure.

Author Response: Hagen Kunte, Felix Schmidt, Jan Hoffmann, Lutz Harms, Golo Kronenberg, Berlin: We appreciate the suggestion that an assessment of olfactory function by the extended Sniffin' Sticks procedure may be helpful in the diagnosis and follow-up of IIH. The prevalence of absolute hyposmia in patients with a recent first diagnosis of IIH or patients with a clinically significant worsening of IIH within the last 3 months reached 80% in our sample.[1] We agree that the presence of olfactory dysfunction should be considered as one of the minor criteria for diagnosing IIH. However, other potential causes of reduced olfactory function must first be ruled out. In contrast, olfactory testing is less appropriate as a follow-up measure in patients with diagnosed IIH because olfactory function typically trails changes in intracranial pressure (ICP) by several weeks. Emerging evidence suggests that 3D spectral-domain optical coherence tomography of the optic nerve head might be used to noninvasively monitor IIH and possibly other disorders with increased ICP.[3]

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2. Schmidt C, Wiener E, Hoffmann J, et al. Structural olfactory nerve changes in patients suffering from idiopathic intracranial hypertension. PLoS One 2012;7:e35221.
3. Kaufhold F, Kadas EM, Schmidt C, et al. Optic nerve head quantification in idiopathic intracranial hypertension by spectral domain OCT. PLoS One 2012;7:e36965.

CORRECTION

Teaching Neuro*Images*: Dyspnea as a presenting manifestation of amyloid myopathy

In regard to the article "Teaching Neuro*Images*: Dyspnea as a presenting manifestation of amyloid myopathy" by P.S. Ghosh et al. (*Neurology*® 2013;81:e184), there is an error in the cover image text found at the end of the Table of Contents. It should have read: "Trichrome stain of a deltoid muscle showing a necrotic muscle fiber in a patient with amyloid myopathy." The authors regret the error.

Author disclosures are available upon request (journal@neurology.org).